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Thrombotic events are a well-recognized complication of homocystinuria. However, the mechanisms involved in the atherogenic and thrombotic effects of homocyst(e) ine remain incompletely understood. The objective of this

study was to determine the role of endothelial cell activation/damage as indicated by levels of thrombomodulin, tissue factor and tissue factor pathway inhibitor, and factor VII activity in patients with homocystinuria. Six

patients with homocystinuria, nonresponsive to pyridoxine, treated only with trimethylglycine (betaine) were injected with a bolus of 20 IU/kg body weight of unfractionated commercial heparin to induce the release of tissue factor pathway inhibitor from the vascular endothelium. Tissue factor, thrombomodulin, and factor VII activity were measured by enzyme-linked

immunosorbent assay and clotting assay before heparin administration. Tissue factor pathway inhibitor antigen and activity were measured before and 5 minutes after the bolus of heparin. Levels of homocyst(e)ine were elevated (patients: 144.2+/-19.2 micromol/L; controls: 10.2+/-0.9

micromol/L); however, levels of thrombomodulin, tissue factor, and tissue

factor pathway inhibitor antigen were not statistically different from the control group. In contrast, tissue factor pathway inhibitor activity showed a significantly increased level (patients: 2.09+/-0.34 U/L; controls: 1.14+/-0.20 U/L; p<0.05) that was correlated with homocyst(e)ine. Factor VII activity was significantly decreased (patients: 64.7+/-5.19/; controls: 01.4+/-4.79/;

was significantly decreased (patients: 64.7+/-5.1%; controls: 91.4+/-4.7%; p<0.05) and inversely correlated with homocyst(e)ine. After heparin the patients released higher amounts of tissue factor pathway inhibitor antigen and activity compared with the control group; however, the difference was not statistically significant. Although not treated with antithrombotic drugs,

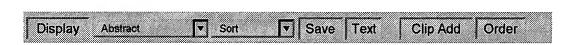
none of the patients had any thromboembolic complications after starting

betaine. In addition to betaine treatment, the enhanced factor pathway inhibitor antigen activity observed in this small series of patients suggests that

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factor pathway inhibitor antigen may play an additional, as yet unexplained, role in this genetic disorder.

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